

JSPCCS-AHA Joint Symposium

JSPCCS-AHA Joint Symposium (I-AHAJS)

Preventive medicine

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[I-AHAJS-04]Effect of a school-based screening program on the outcome of patients with long QT syndrome and hypertrophic cardiomyopathy

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Among major causes of out-of-hospital cardiac arrest (OHCA) in children and adolescents, hypertrophic cardiomyopathy (HCM) and long QT syndrome (LQTS) are leading causes of OHCA. A nation-wide school-based screening program has uncovered patients with these two conditions; however, the effect of the program was different.

In terms of LQTS, among 282 patients with a high probability of LQTS who visited our center between 2005 and 2018, 79% were screened by the program, suggesting that most patients are diagnosed through the program in Japan. The former study of 117 unrelated probands showed that approximately 90% of mutations had radical or high probability of pathogenicity in both the screened and non-screened groups. The study also showed that prevalence of symptoms before and after diagnosis was significantly lower in the screened group when compared with that in the non-screened group although the QTc values were not different between the groups. The data suggest that the program may be effective for early diagnosis and prevention of symptom.

Concerning the HCM, a recent study on childhood cardiomyopathy in Japan found 141 cases with HCM including 38 secondary cases. Of 103 idiopathic cases, 59% were diagnosed by the screening program, and 12 cases had poor prognosis (death, transplantation, or OHCA). Multiple logistic regression analysis showed that the screening program was not a preventive factor for the presence of poor prognosis. Early diagnosis and intervention are needed for patients with childhood HCM.